High prevalence of baffle leaks in adults after atrial switch operations for transposition of the great arteries

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Abstract: Aims To determine the prevalence of baffle leaks in adults after atrial switch operations for transposition of the great arteries, as these may predispose to paradoxical embolic events, particularly in patients with transvenous pacemaker or defibrillator leads. Methods and Results We routinely perform contrast echocardiography with agitated saline in all patients after atrial switch operations. For this study, we analysed patients who had saline contrast echocardiography between 2010 and 2012. The presence of baffle leaks and the severity of right-to-left shunting were assessed. We compared baseline characteristics and oxygen saturation at rest and during exercise between patients with and without baffle leaks. A total of 65 patients (56 Senning and 9 Mustard repair) without previously known baffle leaks were included (mean age 32 ± 8 years, 77% males). Right-to-left shunting was identified in 42 patients (65%) and occurred without provocation manoeuvres in 88%. There were no differences in baseline characteristics, echocardiographic findings, or exercise capacity between patients with and without baffle leaks, except for lower oxygen saturation at peak exercise in those with baffle leaks (29% had oxygen saturations below 90% at peak exercise compared to none without baffle leaks, P = 0.011). Four patients with baffle leaks had previous implantation of transvenous pacemaker leads; one of them had suffered a stroke. Two other patients with baffle leaks had a history of potential embolic stroke. Conclusions Because of the high prevalence of baffle leaks in adults after atrial switch operations, we propose routine screening with agitated saline contrast, particularly prior to implantation of transvenous pacemaker or defibrillator leads.

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High prevalence of baffle leaks in adults after atrial switch operations for transposition of the great arteries

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Conclusions Because of the high prevalence of baffle leaks in adults after atrial switch operations, we propose routine screening with agitated saline contrast, particularly prior to implantation of transvenous pacemaker or defibrillator leads.

Keywords transposition of the great arteries • atrial switch repair • Mustard repair • Senning repair • baffle leaks • stroke

Introduction

Complete transposition of the great arteries is the second most common cyanotic heart defect, affecting about 20–30 of 100 000 liveborns.[1] Without surgical repair, most affected patients die in early childhood.[2] With the advent of open heart surgery and the introduction of the atrial switch repair by Senning and Mustard, the fate of affected patients changed dramatically and most patients survived to adulthood.[3,4] Although the atrial switch repair was superseded by the arterial switch repair in the 1980s at most centres, there is still a large cohort of adults with Senning and Mustard repairs under clinical follow-up. These patients remain at increased risk of cardiovascular complications in adulthood and have a markedly increased mortality risk compared to the general populations.[5] Complications of the surgically created venous baffles (obstruction and/or leaks) substantially contribute to long-term morbidity and predispose to paradoxical systemic embolism. This may be of particular importance, as many patients require cardiac pacemakers and implantable defibrillators for

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sinus node dysfunction or prevention of sudden cardiac death.[6] Studies have shown that patients with intracardiac shunts are at increased risk for paradoxical systemic embolic events in the case of transvenous pacemaker leads.[7,8] Although some studies have investigated the prevalence of baffle leaks in patients who had undergone cardiac catheterization, the overall incidence of baffle leaks among adults after the atrial switch operation is not well defined.[9]

The aim of this study was to determine the prevalence of baffle leaks among adults after atrial switch repair for transposition of the great arteries and to determine their association with clinical and echocardiographic characteristics as well as with previous ischaemic strokes.

Methods

Since 2010 we perform contrast echocardiography with agitated saline contrast as part of our routine follow-up protocol in all adults after atrial switch operations (Senning or Mustard repair) to detect baffle leaks.

Technique of saline contrast injection

For preparation of agitated saline, we use 20 mL Luer lock syringes connected by a three-way stopcock. We mix 16 mL of standard 0.9% saline with 4 mL of air without additional adjunction of patient's blood. If possible, injection of agitated saline contrast is performed via an intravenous line into the right antecubital vein with elevated right arm at the time of contrast injection to facilitate contrast flow to the heart. We first inject only 2–3 mL of agitated saline contrast to avoid massive right-to-left shunting in case of a large baffle leak. If necessary we then repeat saline contrast injection with up to 15 mL agitated saline with the aim to provide dense bubble contrast within the subpulmonic left ventricle. In a second step, we repeat agitated saline contrast injection with Valsalva manoeuvre. In patients with a high suspicion for a baffle leak despite negative saline contrast study after injection into cubital veins, we occasionally perform a saline contrast study with injection into a leg vein, if documentation of a baffle leak had direct clinical implications. To achieve proper bubble-contrast density within the subpulmonic left ventricle after injection into a leg vein, it is important to flush the intravenous line with 20–40 mL of standard saline after the injection of bubble contrast and to elevate the leg to facilitate venous return.

For the purpose of this study, we analysed all patients after atrial switch operation, who had transthoracic echocardiography with agitated saline contrast injection between 2010 and 2012. The presence of baffle leaks and the degree of right-to-left shunting were assessed. In analogy to grading of shunts across patent foramen ovale, the amount of shunting was graded in a semi-quantitative manner (*Figure 1*).[10] Grade 1: few bubbles appear in the subaortic right ventricle after bubble contrast enters the subpulmonic left ventricle; Grade 2: bubble contrast fills large parts of subaortic right ventricle; and Grade 3: Dense bubble contrast within the entire subaortic right ventricle. Furthermore, it was differentiated whether shunting occurred spontaneously or after provocation with Valsalva manoeuvre only.

We compared baseline characteristics, oxygen saturation at rest and during exercise testing, and haemoglobin levels between patients with and without baffle leaks. In addition, the prevalence of implanted pacemakers, previous paradoxical systemic embolic events, and ischaemic strokes were recorded. The study complies with the Declaration of Helsinki and was approved by the local ethics committee.

Statistics

Continuous data are presented as mean ± standard deviation or median with range as appropriate. Categorical data are presented as number with percentage. For comparison between groups, Chi square tests, Fisher's exact test, Student's *t*-test, Mann–Whitney test, or ANOVA tests were used, as appropriate. The two-sided P values < 0.05 were considered statistically significant.

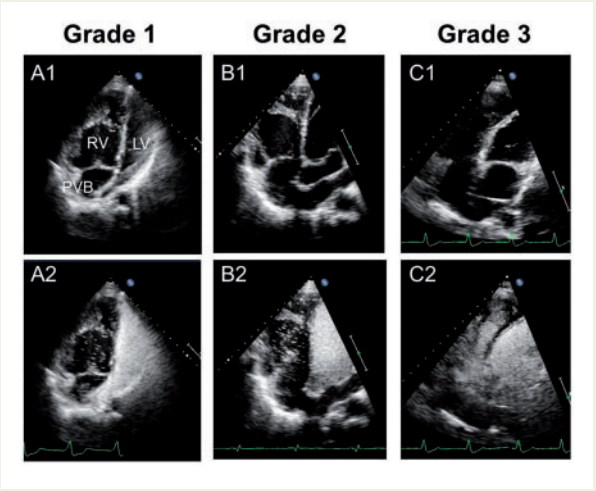

Figure 1 Grading of shunting from systemic venous baffles into pulmonary venous baffles. Upper row, panels A1–C1: still frames of apical four-chamber views prior to bubble-contrast injection. Lower row, panels A2–C2: still frames of apical four-chamber views after bubble-contrast injection. RV, subaortic right ventricle; LV, subpulmonic left ventricle; PVB, pulmonary venous baffle.

Results

A total of 65 patients (56 patients after Senning and 9 patients after Mustard repair) without previously known baffle leaks were identified. Mean age was 31.6 ± 7.9 years, and 77% were males. In 57 patients (88%) saline-contrast echocardiography was performed without clinical suspicion for a baffle leak and in 8 patients (12%) for suspected baffle leaks (four patients had oxygen saturations below 95% in the absence of an alternative explanation, two patients had recurrent strokes, and two patients had suspicion of intracardiac shunts based on findings on cardiac magnetic resonance imaging). Right-to-left shunting (i.e. shunting from the systemic venous atrial baffle to the pulmonary venous atrial baffle and hence from systemic venous to systemic arterial circulation) was identified in 42 patients (65%). Localisation of the exact site of shunting was not possible in most patients and in only two patients shunts could be identified on colour Doppler. Most patients had spontaneous right-to-left shunting (88%) without provocation manoeuvres. Seventeen patients (45%) had Grade 1, 12 patients (33%) Grade 2, and 8 patients (21%) Grade 3 shunts. Two patients with Grade 3 shunts had transient visual disturbances after agitated saline injections. Both episodes were self-limiting within less than 30 min. One of these patients subsequently had cerebral magnetic resonance imaging for a different reason without evidence of ischaemic cerebral lesions. No other adverse events were recorded.

Baseline characteristics of patients with and without baffle leaks are reported in *Table 1*. No differences were found in any baseline

variables between patients with and without baffle leaks. Of all 65 patients, 54 (83%) had been operated at our centre in Zurich and 11 patients had been repaired at other institutions. There was no difference in the rate of baffle leaks between patients who had been operated at our institution or at another institution (67% vs. 64%, $P = 1.0$). Most atrial switch operations at our institution had been performed by an experienced surgical team, including a substantial number of operations performed by the inventor of the surgical technique, Prof. Åke Senning. We found no significant differences of rates of baffle leaks between patients that had been operated by different surgeons ($P > 0.3$ for all comparisons between different surgeons).

Overall, there were no differences in right or left ventricular dimensions or ventricular function between patients with and without baffle leaks (*Table 2*). In 36 patients (55%) data from cardiac magnetic resonance imaging were available. Again, no differences between patients with or without baffle leaks were evident for right ventricular volumes (113 ± 28 vs. 119 ± 24 ml/m^2, $P = 0.30$) or left ventricular volumes (84 ± 25 vs. 87 ± 28 ml/m^2, $P = 0.98$). Five patients with cardiac magnetic resonance imaging had left ventricular end-diastolic volumes indexed to body surface area > 100 mL/m^2; four of them (80%) had a baffle leak. On echocardiography, only four patients had a left ventricular end-diastolic diameter >50 mm on parasternal views, all of these patients had baffle leaks.

Table 1 Baseline characteristics of patients

	Baffle leak (*n* = 42)	No baffle leak (*n* = 23)	*P*-value
Male (%)	33 (79)	17 (74)	1.0
Age (years)	31.4 ± 8.1	32.1 ± 7.6	0.76
Senning repair (%)	37 (86)	19 (86)	0.64
Age at repair (years)	0.9 (0.0-7.6)	0.9 (0.2-8.0)	0.73
Heart surgery before 1984 (%)	19 (45)	14 (61)	0.19
Body mass index (kg/m2)	24.5 ± 5.0	24.7 ± 5.9	0.86
History of atrial arrhythmias (%)	8 (19)	5 (23)	0.75
On oral anticoagulation (%)	11 (26)	6 (27)	1.0
History of previous stroke (%)	4 (9)	1 (5)	0.66
NYHA functional class ≥ II	3 (7)	2 (9)	0.38

Oxygen saturations, exercise testing, and haemoglobin values

While two patients with baffle leaks (5%) had oxygen saturations below 90% at rest (86% and 89%, respectively), none of the patients without a baffle leak had resting oxygen saturations below 90% at rest. On average, there was no statistically significant difference in resting oxygen saturation between patients with and without baffle leaks (97%, range 86–99% vs. 98%, range 94–100%, $P = 0.22$). There was also no statistically significant difference in haemoglobin concentrations between patients with and without baffle leaks (15.5 ± 1.4 vs. 15.4 ± 1.1 g/dL, $P = 0.88$).

In 59 patients (91%), data from exercise testing were available. Although patients with baffle leaks had higher peak heart rate on exercise testing (165 ± 22 beats/min vs. 149 ± 26 beats/min, $P = 0.021$), there was no difference in peak oxygen consumption (25.2 ± 8.3 vs. 24.9 ± 9.2 mL/kg/min, $P = 0.91$) or percent-predicted peak oxygen consumption (69 ± 16% vs. 64 ± 20%, $P = 0.19$) between groups.

While there was no significant difference in resting oxygen saturation, a significant difference in peak oxygen saturation during exercise testing between patients with and without baffle leaks was recorded (*Figure 2*). While none of the patients without a baffle leak had a decrease of oxygen saturation below 90% during exercise testing, 11 patients with baffle leaks (29%) had a minimal oxygen saturation below 90% at peak exercise ($P = 0.011$ for comparison between groups). Desaturation on exercise was more pronounced in patients with larger right-to-left shunts at rest: Patients with a Grade 1 shunt had oxygen saturation at peak exercise of 96.0 ± 2.2%, those with a Grade 2 shunt 92.6 ± 5.0%, and those with a Grade 3 shunt 87.9 ± 5.4 ($P < 0.0001$ for comparison between groups).

Previous ischaemic strokes and pacemakers

Five patients had a history of previous ischaemic strokes, of which one was a peri-interventional stroke at the time of preoperative cardiac catheterization and one patient suffered a perioperative stroke

Table 2 Echocardiographic findings

	Baffle leak (*n* = 42)	No baffle leak (*n* = 23)	*P*-value
Right ventricular end-diastolic area (cm^2)	37.7 ± 9.1	36.7 ± 9.1	0.65
Right ventricular fractional area change (%)	27.2 ± 7.4	29.8 ± 9.9	0.24
Tricuspid annular plane systolic excursion (mm)	13.0 ± 4.6	12.6 ± 3.3	0.76
Right atrial long axis dimension (cm)	5.5 ± 0.9	5.7 ± 1.0	0.37
Right atrial short axis dimension (cm)	3.6 ± 0.8	3.9 ± 0.9	0.16
Left ventricular end-systolic diameter (cm)	2.3 ± 0.9	2.2 ± 0.7	0.56
Left ventricular end-diastolic diameter (cm)	3.8 ± 0.9	3.6 ± 0.7	0.36
More than mild tricuspid regurgitation (%)	9 (21)	5 (23)	1.0

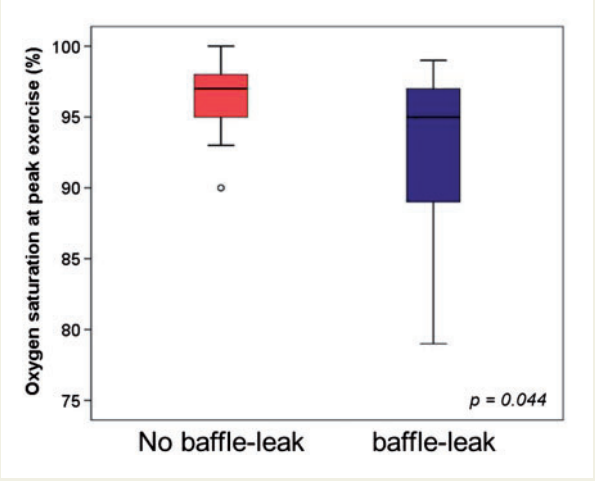

Figure 2 Oxygen saturation at peak exercise.

at the time of intracardiac repair. All three patients with an incidental (non-peri-interventional or perioperative) stroke were found to have baffle leaks with spontaneous right to left shunting, all three had Grade 2 shunts. All three patients were assessed for device closure of the baffle leak, but it was deemed technically impossible in all three cases. All three patients were put on long-term oral anticoagulation therapy with vitamin K antagonists.

Fifteen patients (23%) had previous pacemaker or defibrillator implantation, 8 with epicardial and 7 with endocardial leads. Of patients with endocardial leads, 4/7 (57%) had a baffle leak. One of these four patients had transvenous pacemaker implantation at the age of 9 years for sinus node dysfunction and suffered an ischaemic stroke at the age of 14 years with persistent partial loss of vision. At the time of stroke, no further investigations were undertaken. When he was found to have a baffle leak with spontaneous right-to-left shunting at the time of routine testing (age 34 years), the anatomy of the baffle leak was found to be not amenable to device closure (confirmed at a different institution) and he was put on long-term anticoagulation. One patient with baffle leak and transvenous pacemaker underwent device closure of the baffle leak, which did not lead to complete abolition of right-to-left shunting and thus he was kept on long-term anticoagulation. The other two patients with baffle leaks and transvenous pacemaker leads were put on preventive anticoagulation with vitamin K antagonists, both additionally had atrial arrhythmias and thus a further indication for long-term anticoagulation.

Discussion

Our study demonstrates that baffle leaks after atrial switch operations are very common and affect about two-thirds of patients. In most patients, these baffle leaks cannot be identified on routine Doppler echocardiography, and the majority of patients with baffle leaks did not show any suggestive findings, such as ventricular dilatation or desaturation at rest. In the absence of haemodynamically significant shunts, even cardiac magnetic resonance imaging is an insensitive method for the detection of baffle leaks. While the prevalence of baffle obstructions has been investigated in a number of studies by cardiac magnetic resonance imaging, computed tomography, or cardiac catheterization, the prevalence of baffle leaks in all-comers after atrial switch operations has not been reported. A study published by Wilhelm *et al.*,[9] reviewing patients who had cardiac catheterization for clinical reasons found a prevalence of baffle leaks in 39/76 cardiac catheterization procedures; however, in 24% of these patients, the indication for cardiac catheterization was evaluation of a known baffle leak.

Although most baffle leaks in patients after atrial switch operations are small, their physiology differs fundamentally from inter-atrial shunts in the setting of a patent foramen ovale or a secundum-type atrial septal defect. In patients with a patent foramen ovale, right-to-left shunting is typically present in most cases only after provocation with manoeuvers that increase right atrial pressure (e.g. Valsalva manoeuvre). In contrast, in our study, the majority of patients with baffle leaks had evidence of spontaneous right-to-left shunting with a high proportion having relatively large shunts. This observation may be explained by the fact that systemic venous baffles are often relatively stiff and may increase systemic venous filling pressures and thus promote right-to-left shunting, even in the absence of a defined systemic venous baffle stenosis. Facilitated right-to-left shunting in patients with baffle leaks may indeed predispose to paradoxical systemic embolic events.

If closure of a baffle leak is considered, careful transoesophageal echocardiography to delineate the exact localization, size, and tissue rims of baffle leaks is mandatory. Many patients have more than one leak. Occasionally, three-dimensional transoesophageal echocardiography may provide some additional information that allows better planning of a percutaneous procedure. The technique of baffle leak closure is determined by lesion anatomy and concomitant baffle stenosis. In the absence of concomitant baffle stenosis, we usually use double disc devices for baffle leak closure and reserve covered stents for leaks with concomitant baffle stenosis.

In our cohort, three patients with a baffle leak had a history of incidental ischaemic stroke, one in the setting of previous transvenous pacemaker lead implantation. Given the fact that intracardiac shunts in patients with transvenous pacemaker leads are a well-established risk factor for embolic strokes, the notion that almost a quarter of all patients in our cohort had implanted pacemakers or defibrillators highlights the importance of careful screening for intracardiac shunts before implantation of transvenous devices. Contrast echocardiography with agitated saline contrast offers a cheap, quick, safe, and likely sensitive method for the detection of baffle leaks.

In patients with documented spontaneous right-to-left shunting on saline-contrast echocardiography, the use of air bubble filters for all intravenous lines eliminates the risk of paradoxical air embolism. The liberal use of prophylaxis against venous thrombo-embolism at times of increased risk in order to decrease the likelihood of paradoxical systemic embolism may further decrease morbidity in this vulnerable patient group.

Limitations

As in our routine practice, saline contrast injection is performed from antecubital veins only, the true incidence of baffle leaks may even be slightly underestimated because baffle leaks of the inferior vena cava limb may be missed. In the case of otherwise unexplained dilatation of the subpulmonic left ventricle or oxygen desaturation at rest or during exercise in patients with negative saline contrast studies after

injection in arm veins, it may be necessary to repeat contrast echocardiography with injection of bubble contrast into leg veins. This was not performed routinely in our cohort. None of the patients without neurologic symptoms had cerebral magnetic resonance imaging and therefore the true incidence of paradoxical embolism might be underestimated, as some may be clinically silent.

Although residual baffle leaks may be influenced by surgical technique or expertise of the cardiac surgeon, we found no differences in rates of baffle leaks between patients operated at different institutions or by different cardiac surgeons.

Although our cohort provides some evidence for an increased risk of ischaemic strokes in patients with baffle leaks, particularly in the presence of transvenous pacemaker leads, the small patient number does not allow an estimation of the true risk of paradoxical systemic embolism in patients with baffle leaks.

Conclusion

Given the high prevalence of baffle leaks in patients after atrial switch operations, we propose routine screening with agitated saline contrast, particularly prior to implantation of transvenous pacemaker or defibrillator leads.

Conflict of interest: None declared.

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